

A Rare Case Report on Non-Functioning Benign Spindle Cell Neoplasm- Adrenal Schwannoma

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ABSTRACT

Schwannoma or benign tumor of the myelin sheath which are rare and can be functioning or non-functioning that are difficult to diagnose preoperatively. Visceral schwannoma in the adrenal region is extremely rare, accounting for only 0.7% of the adrenal tumors. Here we report such a rare case of Adrenal schwannoma in 40 years old female patient with hypothyroidism.

KEYWORDS: Adrenal adenomas, Adrenal tumors, Schwannoma, benign spindle cell neoplasm, Adrenal gland

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INTRODUCTION

Adenomas of the adrenal gland are non-cancerous tumors on the adrenal gland. Most of them don't cause any signs or symptoms. However some may become active /functioning which means they produce hormones. Often in excess of what adrenal glands typically produce. Highly levels of these hormones can lead to complications including primary aldosteronism, Cushing's syndrome and many other medical conditions. Majority of adrenal adenomas are Non-Functioning, they don't produce hormones and usually don't cause any symptoms. They often found incidentally. Schwannoma also known as Neurilmmoma which are rare benign tumors with very low incidence of malignant changes arising from Schwann cells of peripheral, motor, sympathetic or cranial nerves. The most common site of origin is upper and lower extremities, head and neck. They are also found in juxta-adrenal and retroperitoneum areas. Schwannomas presenting in visceral organs especially the adrenal gland are rare [1-7]. We report such a rare case in 40 years old female patient.

CASE PRESENTATION

A 40 year old female patient visited our hospital with chief complaints of pain at left lumbar region since 4months. She

is a k/c/o hypothyroidism since 5 years. A general physical examination was unremarkable with blood pressure of 130/85mmhg; pulse rate 80 beats/min and sinus rhythm. Blood picture and biochemical profile were within normal range. Serum FT3-4.07pmol/L, Serum TSH-7.77µIU/mL, Serum FT4-16.44pmol/L. Metanephrine- free-8.87pg/mL X-ray chest PA view revealed normal. Ultrasound examination: showed hypoechoic mass 43/40 mm in the left adrenal gland.



Fig.1 showing hypoechoic mass in the left adrenal gland

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CT Abdomen (plain & contrast) reveals well defined hypodense lesion (AVG 11 HU) with speck of calcification seen arising from lateral limb of left adrenal gland. Medial limb and body appear normal. On contrast administration the lesion shows minimal enhancement on arterial phase. On venous and delayed phases there is significant enhancement at the periphery with non enhancing center-likely necrosis. No filtration into adjacent structure seen- possibility of Adrenal Adenoma / Pheochromocytoma to be considered. Polypoidal lesion showing significant enhancement arising from posterior wall of pre pyloric region of stomach. No extension beyond the wall seen.

Histopathology report: On gross examination left supra renal mass measuring 0.4cm is observed. On microscopic examination linear cores revealed hypo cellular areas with hemorrhage spindled cells with occasional degenerating cells and edge of the cores revealed dilated spaces lined by cuboidal cells. HPR report reveals predominantly stroma with blood and few randomly scattered spindle cells. Possibility of mesenchymal lesion- Benign Spindle cell Neoplasm-Schwannoma.

Patient was planned for laproscopic adrenalectomy.

DISCUSSION

Schwannoma is a neurogenic tumor, generally occurring between the third and the sixth decades of life, with no sex or location predilection. In fact, it may arise everywhere in organs or nerve trunks and the with sole exception of cranial nerves I and II, lacking Schwann cells. Firstly Schwannomas were described by Verocay in 1908 and Antonini in 1920 subclassified these tumors into two distinct histologic patterns. Typical Schwannoma is a solitary mass, with ovoid or spherical morphology and well-defined margins; when Schwannoma is very large (>8–10 cm) it is frequently affected by degenerative changes with cystic areas, calcifications, interstitial fibrosis and hyalinization. The degenerative pattern is predominant in the “ancient” Schwannoma, a variant with a very good clinical outcome. It should be known that 5–18%

Of Schwannomas are associated with von Recklinghausen’s disease; in this pathological context Schwannomas are generally malignant with a trend of manifestation in multiple areas [8–10]. The medulla of adrenal gland receives double innervations by two different groups of myelinated nerve fibers, respectively derived from the sympathetic trunk (or vagus nerve) and from the phrenic nerve. Adrenal Schwannomas are thought to origin from one of those nerves and to arise from medulla because of the uninterrupted continuance between the tumor and the adrenal medulla in absence of septum around the tumor. They grow up from adrenomedullary site compressing the adrenal cortex^[1].

The differential diagnosis of an adrenal schwannoma depends on whether it exhibits conventional or cellular histologic features. The diagnosis of conventional schwannomas can typically be made based on their distinctive histologic

features that are described above, such as the alternating Antoni A and B areas, nuclear palisading, and Verocay bodies. Immunohistochemistry with S100 is used mainly for confirmation of the diagnosis. Cellular schwannomas, consisting almost entirely of intersecting fascicles of spindle cells, on the other hand, can present more of a problem in that they can mimic many other lesions with similar morphology. Pheochromocytomas do not uncommonly present as spindle cell lesions. However, they typically display a nested architecture, unlike schwannomas. In addition, they are almost always positive for synaptophysin and chromogranin, unlike schwannomas as described above. Also, pheochromocytomas are negative for S100. Another adrenal tumor that presents with intersecting fascicles of spindle cells is a ganglioneuroma. Although ganglioneuromas contain ganglion cells, there may be areas in the tumor where there is a paucity of these cells, giving an appearance of lesion composed entirely of spindle cells. Distinguishing the adrenal schwannoma from this tumor involves extensive sampling to ensure no ganglion cells are present, as this would be definite evidence for the presence of a ganglioneuroma.

CONCLUSION

We report on an adrenal schwannoma in a 40-year-old female patient. Pre-operative diagnosis of these schwannomas is difficult. Absolute diagnosis is only possible by surgically removing the tumor, and histopathological and immunohistochemical staining of the operative specimen.

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